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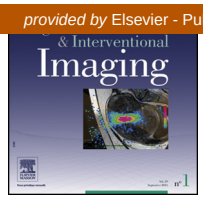

LETTER / *Genito-urinary imaging*

Aggressive angiomyxoma of the pelvis with inferior vena cava involvement: MR imaging features

Keywords: Aggressive angiomyxoma; Tumor thrombus; Inferior vena cava; MRI

Case report

A 50-year-old woman with past history of aggressive angiomyxoma diagnosed in 2002, located initially in the right ovary and treated by hysterectomy and bilateral salpingo-oophorectomy was referred for lower limb edema. She had two pelvic recurrences in 2003 and then in 2004, which were treated surgically and had not been offered any specific follow-up since the last recurrence.

In March 2013, she developed pain and edema in her legs and underwent lower limb venous Doppler ultrasound which showed thrombosis in the right common iliac vein extending into the inferior vena cava. As the thrombosis had not improved despite effective dose anticoagulation for 15 days, imaging was proposed.

An abdominal and pelvic CT without and with iodine enhancement (70 seconds and 3 minutes) showed a bulky pelvic mass on the right side in contact with the iliac vessels measuring $12.5 \times 10 \times 9$ cm, and extending from the ischiorectal fosse to the sacral promontory, pushing the bladder and right ureter anteriorly. Contrast enhancement was late and heterogeneous. The thrombus was of reduced density on all images, without enhancement at all phases of the investigation (Fig. 1) and extended from the right iliac vein through the inferior vena cava into the right atrium. The distinction between tumor thrombus and a fibrin thrombus could not be made from CT findings alone and an abdominal and pelvic MRI with gadolinium, centered on the inferior vena cava was proposed in order to establish that the thrombus was made up of tissue. The investigation included T1 and T2 weighted images without fat saturation in the axial and coronal planes and 3D echo gradient T1 weighted images with fat saturation, both without and with gadolinium chelate (2, 3, 5, 10 and 17 minutes) in the same planes. The pelvic tumor was slightly heterogeneous and hypointense on the T1 weighted images, and heterogeneous and hyperintense on the T2 weighted images with a ''layered'' appearance (Fig. 2). The pelvic tumor enhancement was heterogeneous, pseudo-layered and seen

Figure 1. Enhanced chest and abdominal CT scan in the portal phase, thin MIP (maximum intensity projection) coronal reconstruction. The thrombus extends into the inferior vena cava and the right atrium (white arrow) while the pelvic mass is in contact with iliac vessels (white star).

3 minutes after gadolinium injection. The thrombus was hypointense on the T1 weighted images, and hyperintense on the T2 weighted images compared to the muscles and isointense on T2 compared to the tissue component in the pelvic mass. Local contrast uptake was found late within the thrombus at 10 minutes and was seen better at 17 minutes after gadolinium injection (Fig. 3).

The patient initially underwent thrombectomy through a sternotomy and right atriotomy under extracorporeal circulation in order to reduce the risk of thromboembolism. The surgical specimen contained only tissue which was not adhering to the inferior vena cava wall and was extracted simply by traction (Fig. 4).

In a second operation stage, the pelvic mass was excised and the right internal iliac vein was then ligated. The patient was offered but refused medical treatment with an LHRH agonist.

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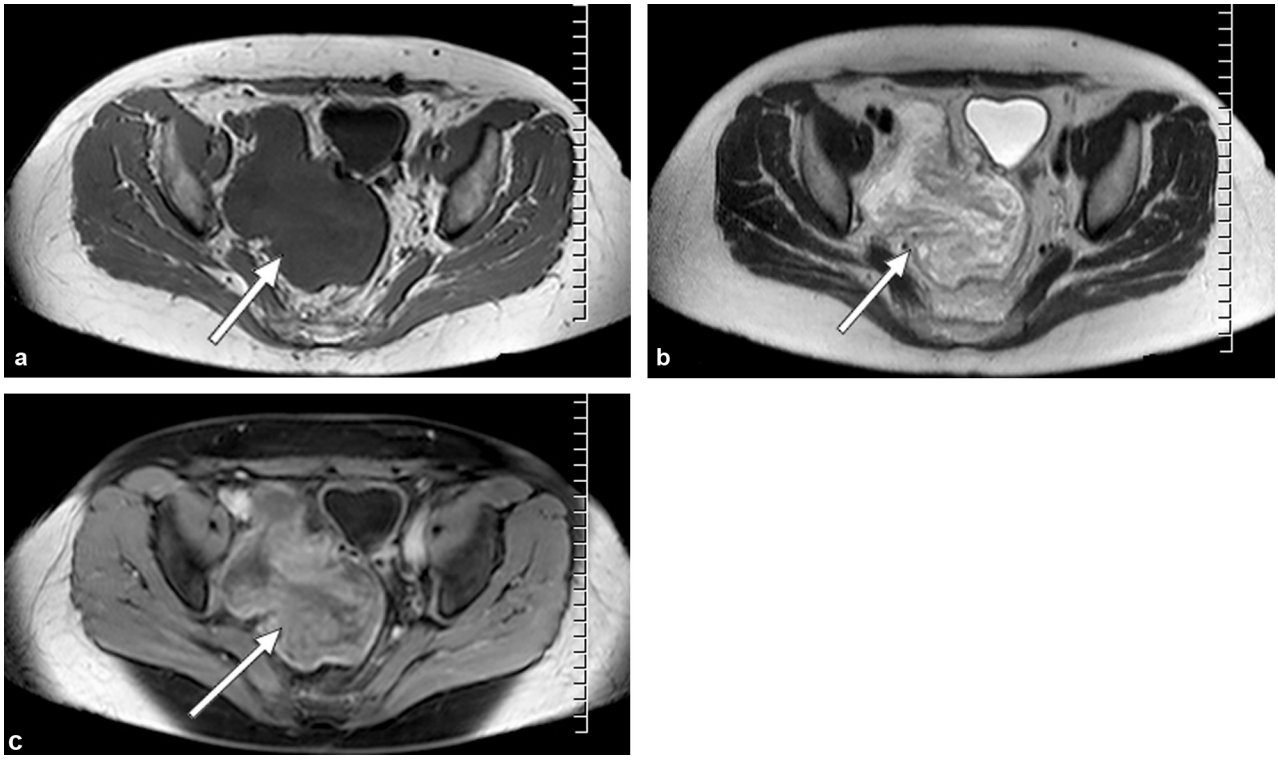

Figure 2. Pelvic MRI. Aggressive angiomyxoma characterization. a: unenhanced axial T1 weighted image without fat suppresion. The tumor is hypointense and heterogenous relative to muscles (white arrow); b: axial unenhanced T2 weighted image without fat suppresion. Heterogenous hyperintense signal of the tumor with swirling or layering appearence (white arrow); c: axial enhanced T1 weighted image with fat suppresion, 3 minutes after the injection of gadolinium. Late, heterogenous enhancement of the tumor with a persitant layering appearance (white arrow).

Histopathological analysis showed both fibrous, myxoid and vascular tumor proliferation containing relatively non-atypical round or oval cells with no mitoses, which on immunohistochemistry labelled strongly for estrogen and progesterone receptors confirming a diagnosis of tumor thrombus secondary to a further recurrence of the aggressive pelvic angiomyxoma.

Discussion

Angiomyxoma is usually a benign tumor that displaces neighboring organs rather than invading them. It is more common in women and mostly develops in the pelvis, vulva or perineum [1,2]. In men, it may develop in the perineum or inguino-scrotal region [3]. Rarer sites have been described, including the pulmonary artery [4]. The aggressive nature of the tumor is due to its potential to infiltrate pelvic organs and the high risk of postoperative recurrence [2] which was estimated to be 47% with the meta-analysis reported by Chan et al. on 106 patients, 71% of recurrences occurring during the first three years [5]. Metastases are rare, although secondary lymph node, peritoneal and lung sites have been described [6,9]. In our case, the presentation of the pelvic tumor on CT and MRI was consistent with findings reported in the literature. The pseudo-layered late enhancement was due to the fibrous component in the tumor, whereas the myxoid and fluid component is reflected by a heterogeneous layered or whirl hyperintensity on T2 weighted images [7].

Invasion of the inferior vena cava, however, is extremely rare and to our knowledge only two cases have been reported. The first was a 38-year-old woman who developed inferior vena cava thrombus extending into the right cardiac cavities which could not be shown to be tissue on transesophageal echocardiography or CT but only on biopsy [8]. The second was a third pelvic recurrence in a 38-year-old woman with extension into the inferior vena cava associated with a lung metastasis [9]. CT showed extensive punctiform contrast enhancement in the thrombus which may, according to the authors, have been due to arterial venous fistulae. Neither of these patients had MRI. In our case, MRI suggested that the thrombus was made up of tissue as a result of the relatively late 3D T1 weighted appearances at 10 and 17 minutes after gadolinium, showing focal punctiform areas of enhancement. The enhancement was difficult to identify as the thrombus was paucicellular on histology, compared to the predominant myxoid component. This is an amorphous substance which does not enhance. Histological examination of the pelvic tumor showed no histological differences with the vena cava thrombosis and in particular the proportion of fibrous tissues and vessels were equivalent. There was, however, a greater proportion of myxoid material in the thrombus compared to the pelvic tumor. This finding may explain the difference in enhancement kinetics between the pelvic tumor and thrombus. It was also impossible to demonstrate on imaging whether or not the thrombus was adhering to the wall. Brunelle et al. described a case of an aggressive pelvic angiomyxoma in a 51-year-old female patient in whom a multi-modal examination was performed. The tumor was hyperintense on DWI images at b0 and b1000 with a high

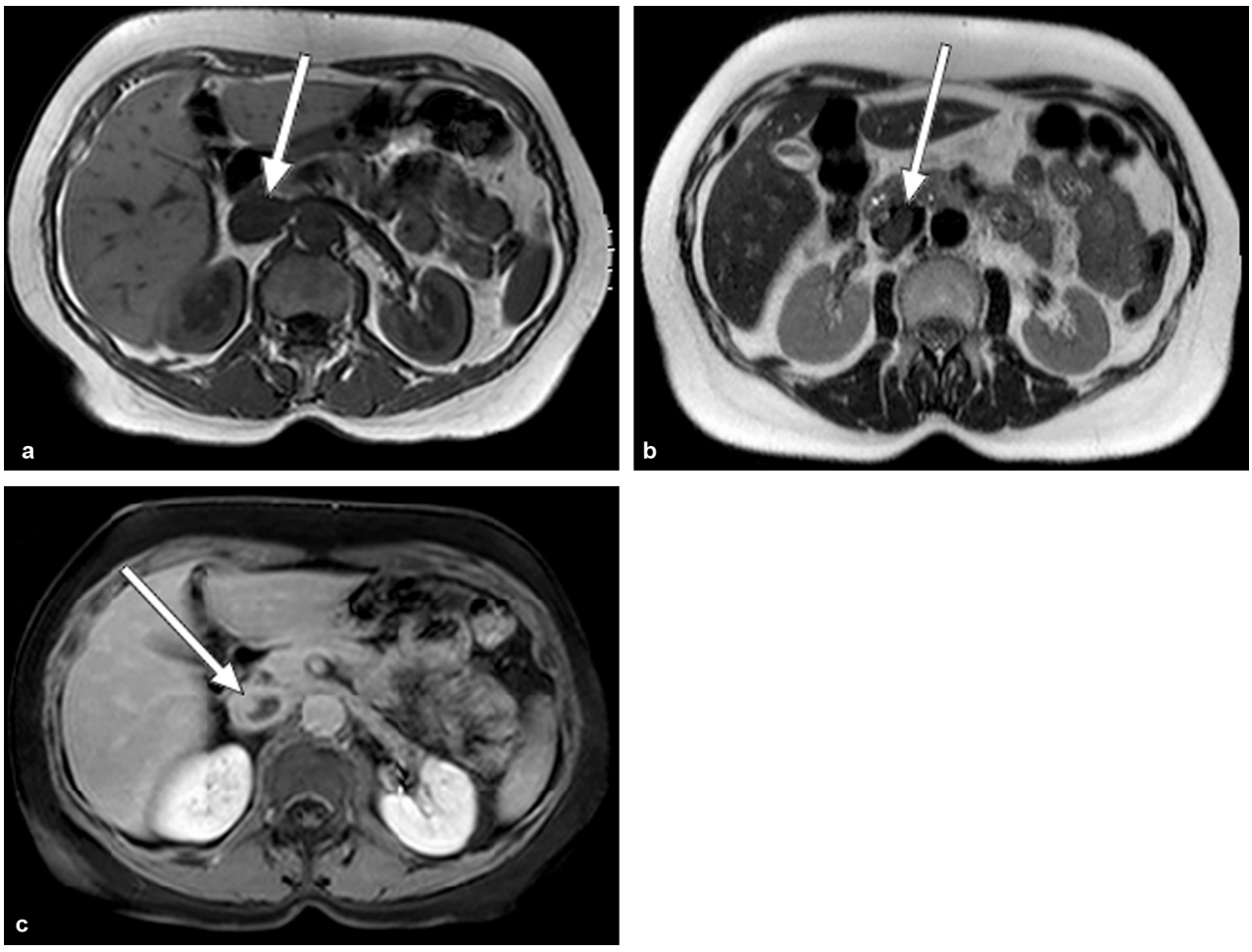

Figure 3. Abdominal MRI. Characterization of the inferior vena caval thrombus. a: unenhanced axial T1 weighted image without fat suppresion. The thrombus is hypointense on the T1 weighted image (white arrow); b: axial unenhanced T2 weighted image without fat suppresion. The thrombus is hyperintense on the T2 weighted image (white arrow); c: axial enhanced T1 weighted image with fat suppresion 17 minutes after the injection of gadolinium. Late punctiform enhancement of the thrombus (white arrow).

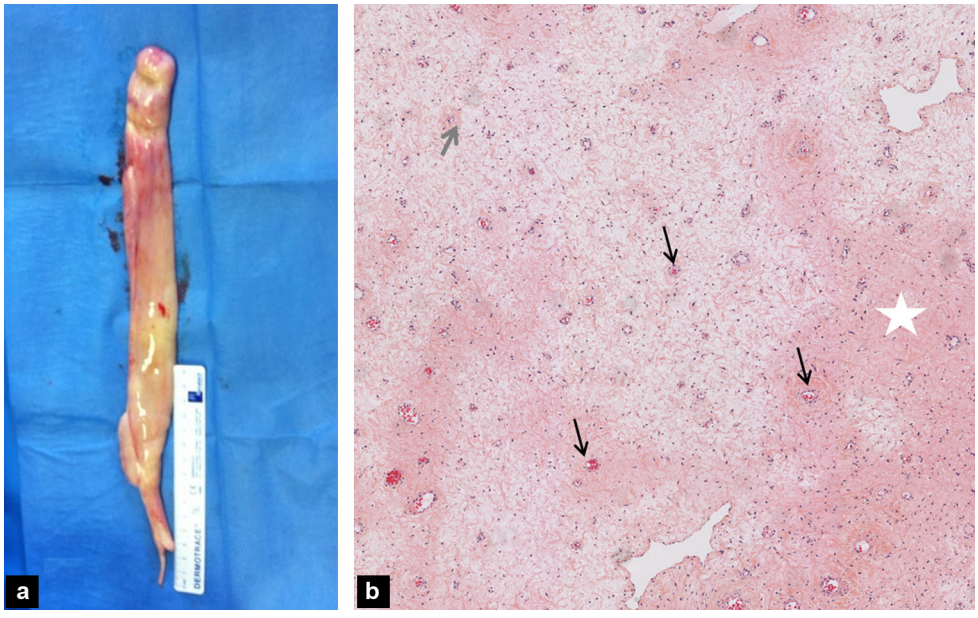

Figure 4. Macroscopic and microscopic aspects of the thrombus. a: photograph of the surgical specimen. The thrombus is made up exclusively of tissue, moulding the inferior vena cava; b: histological section with HES staining. The thrombus is made up exclusively of low cellularity tissue and a large number of vessels (black arrows), collagenous fibers (white star) and myxoid material (gray arrow).

ADC of $2 \times 10{-}3\,mm^2/sec$, similar to what is found in myxoid tumors [10]. The dynamic enhancement examination of this tumor after gadolinium showed gradual enhancement with no plateau or washout, suggesting that it was fibrous in nature.

In terms of treatment, apart from wide surgical excision, an LHRH agonist in a hormone-dependent aggressive angiomyxoma may reduce tumor volume but does not reduce the risk of recurrence [2]. Our patient has not had any further recurrences to date, almost one year after the episode described in this article.

Conclusion

Aggressive angiomyxoma is a histological benign tumor which does, however, have considerable potential to infiltrate neighboring organs and carries a high risk of postoperative recurrence. MRI follow up within 3 years after initial surgery is required in order not to miss a bulky recurrence, which may remain asymptomatic for long periods of time. New treatments with LHRH agonists appear to be promising because of the hormone-dependency of aggressive angiomyxoma, in both reducing tumor volume and limiting mutilating surgery.

Endovascular extension of the tumor is extremely rare and it was only possible to confirm that the thrombus consisted of tumor by MRI, using late 3D echo gradient T1 weighted images over 10 minutes after gadolinium injection. This diagnosis enabled optimal vascular and oncologic surgery to be planned.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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